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Valve-sparing operation in a young woman with Marfan syndrome: A word of caution

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Valve-sparing operation of the aortic root is frequently proposed to young patients with Marfan syndrome as an alternative to aortic root replacement, notwithstanding the complex problems and complications with the remaining aorta. We report the case of a young woman with Marfan syndrome who, 1 year after having undergone a Yacoub procedure, became pregnant and had a recurrence of the aortic insufficiency (AI).

Clinical Summary

A 27-year-old woman with Marfan syndrome underwent a Yacoub procedure for an aneurysm of the aortic root (diameter, 53 mm) with a grade 1+ AI (Figure 1). At the operation, the aortic valve was biscupid and the aortic annulus measured 31 mm. The Yacoub procedure was performed with a 26-mm Dacron tube after resection of the entire aortic wall, leaving 2 mm necessary to perform the suture of the neosinus. The postoperative echocardiogram showed a grade 1+ AI, an aortic annulus diameter of 30 mm, and a neosinus diameter of 43 mm. The patient, while being treated with β-blockers, became pregnant 1 year later and presented during the pregnancy with progressive aggravation of the AI (grade 3+) without hypertension. Two days after cesarean section she had an acute dissection of the descending aorta, which was treated medically. The AI was due to a localized dilatation of the residual aorta at the level of the neosinus, measured on the computed tomographic scan at 47 mm (Figure 2, *A* and *B*). A 2-stage operation was planned to treat both the AI and the dilated descending aorta (dilatation of the descending aorta: 15 mm in 18 months).

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A mechanical aortic valve (size 27 mm, Bicarbon; Sorin Biomedica Cardio S.p.A, Saluggia, Italy) was implanted through an aortotomy in the Dacron tube. The bicuspid aortic valve had a dilated annulus and a prolapsed noncoronary leaflet. The aortic root dilatation was plicated and excluded with stitches passed through the aortic annulus toward the inferior aspect of the Dacron tube. This procedure was performed in association with an elephant trunk procedure with the additional goal to perform a thoracoabdominal replacement (Figure 2, *C*). Aortic crossclamp and pump times were 103 and 139 minutes, respectively. The patient was discharged on postoperative day 6 to a postoperative heart rehabilitation center.

Discussion

The Bentall procedure is usually recommended for patients with Marfan syndrome who have ascending aortic aneurysms.[1] However, because of potential mechanical valve–related complications

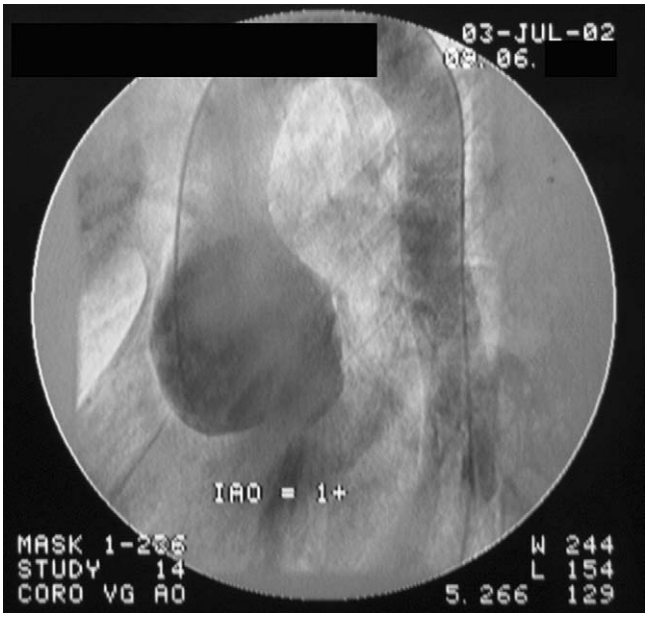

Figure 1. Preoperative angiogram showing dilated aortic root and grade 1+ AI.

Figure 2. Thoracic computed tomographic scan showing localized dilatation of the residual aorta at the level of the neosinus, from the Yacoub operation, measured at 47 mm (A). Reconstruction showing ascension of the reimplanted coronary ostia because of the neosinus dilation (B). Postoperative computed tomographic scan showing the surgical result (C).

such as thromboembolism, hemorrhage, or infection, valve-sparing operations have been proposed in younger patients. These operations, either remodeling or reimplantation,[2,3] have recently regained interest because they preserve the dynamic structure and function of the aortic valve and avoid the need for long-term anticoagulation. Furthermore, they potentially simplify the management of pregnancy, with the lack of anticoagulant therapy. The Yacoub procedure was reported as feasible in more than 70% of patients with Marfan syndrome with good early and long-term results.[3] These results compared favorably with those reported for the Bentall procedure,[1] although no strict comparison has been made.

Our patient had satisfactory initial results with the Yacoub procedure. However, complications probably occurred because of the combined effects of pregnancy on the remaining aortic wall, with the histologic abnormalities of the aorta seen in the case of a bicuspid aortic valve.[4] Classically, pregnancy is a high-risk period for women affected by Marfan syndrome.[5] However, a number of patients having undergone aortic root replacement have had successful pregnancies without complications,[5] providing the patients had echocardiographic monitoring and β-blocker treatment, as in this case. In this context with a young woman, a valve-sparing operation that necessitates leaving some aortic wall remains questionable, particularly in the case of a bicuspid aortic valve. A more radical resection of the ascending aorta should be balanced with a valve-sparing operation, which has the great advantage of not requiring anticoagulant therapy during pregnancy. Another approach in valve-sparing operations is the Tirone David reimplantation technique with a cylindrical tube graft.[2] This potentially limits the increase in the surface area of the leaflet caused by the root dilatation, owing to the annulus inclusion. However, as the absence of sinuses of Valsalva is supposed to perturb the coronary flow and to create an abnormal leaflet stress, which theoretically could limit the long-term durability of the valve, the original technique has led to many variants. An operation combining the advantages of both approaches by adding an external subvalvular prosthetic ring annuloplasty to the remodeling procedure could be a good option in preventing such dilatation[6] and could have been discussed with a more radical operation such as the Bentall procedure in this case.

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